

Personal Experiences in Heart Disease in Childhood

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It is with great appreciation that I have accepted your kind invitation to deliver the Menary Lecture here in Belfast today. It was suggested that I speak of my experiences in the field of heart disease in childhood. There is here, I am sure, no need for me to review the historical aspects of the subject prior to the beginning of the twentieth century. It was in the old world that almost all medical advances in this field took place, until recent years, and since the turn of the century our labours in the States have been largely supplemental, and happily, on occasion, co-operative.

I shall present to you my own experiences in the field, starting, as a matter of fact, with my own boyhood, when soon after the turn of the century I was terribly distressed by the serious illness and death of my small sister, Dorothy, from fulminating recurrent rheumatic fever with pancarditis. Her death, at the early age of 12, was one of the reasons why I took a special interest both in pædiatrics and in heart disease. It was soon after her death that I decided to study medicine, and later on, during my internships at the Massachusetts General Hospital, that I decided to enter the field of pædiatrics. I was somewhat deflected from this early decision by an opportunity to study for a year in London in cardiac physiology and electrocardiography, at the University College Hospital, with Thomas Lewis. It was during that same year of 1913 to 1914 that I had the great privilege of visiting, on occasion, the clinic of Sir James Mackenzie at the London Hospital, and to become acquainted with John Parkinson, who was then Mackenzie's right-hand man, and who has remained a close friend of mine ever since.

For many years I was privileged to direct the Children's Heart Clinic at the Massachusetts General Hospital, and to visit, on occasion, with my colleagues at the House of the Good Samaritan in Boston. In those earlier years our main interest in children was, of course, with rheumatic heart disease. We did recognize a few congenital heart patients, but we knew very little about this difficult

*The Menary Lecture in the Department of Child Health was founded under the will of Mrs. A. J. C. Menary. The holder is required to deliver a lecture on some clinical aspect of child health.

subject then, and we could do even less for the patients. Important aspects of the Children's Heart Clinic at the Massachusetts General Hospital just after the first World War and through the 1920's and 1930's included first, the magnificent work accomplished by a Women's Committee for the Home Care of Children with Heart Disease, second, the Social Service planning of Miss Ida Cannon, who was a sister of Walter Cannon, former Professor of Physiology at Harvard, and also first assistant to Richard Cabot, who established hospital social service, as well as the famous CPC records and clinical ministry at the Massachusetts General Hospital, and third, an early appointee as Social Service Chief of this Children's Cardiac Clinic, Miss Edith Terry, who for many years sparked several pioneer projects for the children, and for the families of our small patients. One of the most interesting and helpful techniques ever devised for both spiritual and physical health of these children, was the In-Bed Club with its jacket, magazine, and visiting and school teaching programmes. In fact, this was so successful that it was finally decided that there should also be an Out-of-Bed Club into which these children would like to graduate from the In-Bed Club. The idea spread through the country and allied chapters were established in other cities. I shall never forget how, on one occasion through occupational therapy, one of our small patients, a boy of 10 or 11, acted as the sole contributor to family funds, while in bed, by making belts and purses, during the illness of both his father and his mother. This gave to him, and the whole family, great satisfaction, as one can well imagine. Fortunately, there is now less need for this Committee of women, and for our social workers, due to a decrease in the severity and the amount of rheumatic heart disease in our midst in New England.

On the other hand, the problem of congenital heart disease has been increasing, so that there is still a great challenge of heart disease in childhood. For the next generation, at least, we shall still have to contend with it, I am sure, despite all our advances in treating active rheumatism, in surgery for mitral stenosis, aortic valve deformity and congenital defects which will challenge the best of our surgeons, and in epidemiology. We are beginning to accept the challenge of the study and prevention of the fundamental factors in these diseases. This last challenge will need also the attention of human geneticists of which we have far too few today.

I have just mentioned the fact that rheumatic heart disease has seemed to be on the down-grade while congenital heart disease has become more of a problem. There are some statistical errors here to which reference should be made. It is, I am sure, quite true that the rheumatic problem is decreasing. For example, we no longer have a long waiting list at the House of the Good Samaritan; in fact, there have been empty beds there of late years. On the other hand, there has not been an actual increase in congenital heart disease. We have simply become more able to diagnose the various defects, some of which used to be called rheumatic (for example, congenital aortic stenosis). At least as important, I suppose, is the fact that these children born with cardiac anomalies, such as occur in blue babies, used to remain in their home communities because there

was nothing which could be done for them in treatment, but now they have, in the last decade, flocked to the medical centres where they have been diagnosed, and often improved or cured by surgery. This explains, I am sure, the increase that we found statistically in the percentage of congenital cardiac patients among our total cardiac population in New England in the course of 25 to 30 years (White 1953). In the 1920's congenital heart disease comprised only 1.5 per cent. of all our cardiac patients, while in the 1950's it had risen to 7.9 per cent. Meanwhile, in that same period, rheumatic heart disease was decreasing from about 33 per cent. to 22 per cent.

RARE KINDS OF HEART DISEASE IN CHILDHOOD.

Now let me discuss in more detail these two particular kinds of heart disease, that is, rheumatic heart disease and congenital heart disease, and my experience with them. There are, to be sure, other kinds of heart disease besides these two varieties, even in childhood, but they are relatively uncommon, and some of them have been almost wiped out. For example, diphtheritic heart disease, which used to kill a generation ago and which could still kill if diphtheria were common, is now rare. Death came in the past by the destructive effect of the diphtheria toxin on the myocardium itself. Also, co-called "congenital hypertrophy of the heart," which used to be an occasional finding, is now rare as such, because it has been, for the most part, subdivided into several minor categories, for example, glycogen storage disease (von Gierke's disease), endomyocardial fibroelastosis, and rare instances of the effect of virus diseases such as mumps which can cause a-v and bundle branch block. I recall very well looking, many years ago, at infants' hearts which showed, with or without congenital defects, a markedly thickened and whitened endocardium, wondering what in the world caused it. This is now generally classified as endomyocardial fibroelastosis. It is not limited to childhood. I myself have encountered two older adults, one in the fifties and one in the seventies, with this condition which I don't think was congenital in their cases. One of these patients, under my observation for about twenty years, finally died of congestive failure after years of coronary insufficiency with bundle branch block. The coronary arteries were found to be but little affected at autopsy, but marked endomyocardial fibroelastosis was present at his age of 74. This condition is of unknown cause; it used to be ascribed to foetal endocarditis, but of late it has been thought more likely due to chronic ischemia.

And now to discuss in more detail the major types of heart disease in childhood, namely, the rheumatic and the congenital.

RHEUMATIC HEART DISEASE.

Our knowledge of rheumatic heart disease has developed considerably during the past generation. One interesting evolution of old thoughts about its relationship to bacteria is the confirmation, on the basis of well established fact, that the hemolytic streptococcus is primarily responsible, producing a reaction of the collagen tissues of the body to its by-products. A long chain of observers, from the last years of the nineteenth century right up to the time of Coburn's findings in the twenties, have presented an interesting chapter in medical history.

There still remains, however, the puzzle of the chain of reaction from the time of the implantation of the streptococcus to the onset of rheumatic fever, an interval that is often quite clearly limited to a period of ten days to a fortnight. Whoever discovers the immunological and biochemical evolution in this chain may thereby afford us the opportunity to break that chain and to prevent rheumatic fever in the relatively small percentage of individuals who are candidates for that disease.

A second interesting finding in the last generation has been that of the familial inheritance of susceptibility to the disease. This has been found to vary from one-third to two-thirds of the patients studied. Probably about 50 per cent. is a reasonable average in the studies reported, that is, about 50 per cent. of the families of patients with rheumatic fever or rheumatic heart disease have in their membership other individuals similarly affected over one or two generations. It is, therefore, as important for us of the medical profession to spend as much time in recognizing the candidates for rheumatic fever as in defining and applying protective and preventive measures. Hence, one cannot be too careful in the study of such a patient in obtaining accurate family histories. So far as possible one should include examination of other members of the family. In this connection I have recently suggested that it would be well, not only for practising physicians to take more complete and adequate family histories, but also that families should keep better records of their own health and longevity. Such a procedure was common a generation or two ago in New England, and, I dare say, in Old England too. Blank pages, properly labelled to record births, marriages, deaths, and other family events, were bound in the midst of the family Bible. The return to such a practice could be very useful for our descendants, whether incorporated in the family Bible or not. When I spoke of this recently, someone suggested that it might be worth while to revive the family Bible itself. Perhaps they both could be revived together.

In the 1930's Drs. Duckett Jones, Edward Bland, and I came to realize, from our study of the youngsters both in the Children's Heart Clinic at the Massachusetts General Hospital and in the House of the Good Samaritan in Boston, that many of the signs, including cardiac enlargement and murmurs of various sorts, could readily come and go when the heart dilated under the stress of rheumatic activity. This was not infrequently found then, and still can be found in cases of severe rheumatic myocardial disease, often with pancarditis. It is especially interesting to find that even mitral diastolic murmurs, which we used to think were diagnostic of mitral stenosis or attributable to the effect of aortic regurgitation demonstrated by Austin Flint, could be due to temporary dilatation of the left ventricle, often lasting for weeks or months during the acute rheumatic attack. On occasion with recovery these murmurs, both mitral systolic and mitral diastolic, and even in a few instances, aortic diastolic, would disappear. There once were controversies about such findings, but now I am sure we all recognize the firm establishment of this possibility. Perhaps one of the most important follow-up studies ever made has been that of the cases at the House of the Good Samaritan by Bland and Jones. When I was in Moscow last September, I found that even there this particular follow-up study was considered as the basis for an hour's

teaching. It is, in fact, so important that I would like to quote now from the Summary and Conclusions of the last reports by Bland and Jones (1951).

“From a twenty-year study of 1,000 patients with rheumatic fever and/or chorea, followed since childhood, the major events of the two decades have been summarized and compared with the experience of others.

“On recovery from the initial illness, 653 patients had signs of rheumatic heart disease. By the end of twenty years the signs of heart disease had disappeared in 108 (16 per cent).

“The remaining 347 patients recovered from their initial illness without detectable heart disease (potential rheumatic heart disease). By the end of twenty years 154 (44 per cent.) had acquired signs of valvular disease.

“During the first ten years 202 succumbed, and by the end of the second ten years 301 had died. Rheumatic fever and congestive heart failure accounted for 80 per cent. of the fatalities, and bacterial endocarditis for an additional 10 per cent.

“A greatly enlarged heart or congestive failure early in the disease exacted the highest toll, with an 80 per cent. mortality in twenty years. Pericarditis, subcutaneous nodules, and acute arthritis occupied intermediate positions, with 63, 37, and 27 per cent. mortality, respectively, in two decades. In contrast, chorea was associated characteristically with a benign form of the disease (12 per cent. mortality).

“Recurrence of rheumatic fever or chorea occurred in approximately one in five during the first five years, one in 10 during the next five years, one in 20 during the third five year interval, and much less frequently in the final five year period.

“A pure form of mitral stenosis evolved in 117 patients, but in only 12 has evidence of serious pulmonary hypertension appeared (acute pulmonary oedema).

“It is encouraging that three out of four of the 699 survivors have little or no limitation.”

One of the most puzzling of all the problems has been that of trying to establish criteria for the activity of the rheumatic process. A well known paper of the late Duckett Jones, published in 1944, is also worth quoting very briefly :—

“For the present it would seem advisable to limit the diagnosis of rheumatic fever to patients with rather distinct clinical manifestations. It is suggested that the following constitute reasonably certain diagnostic criteria :—

1. Any combination of the major manifestations (carditis, arthralgia, chorea, nodules and a verified history of previous rheumatic fever).
2. The combination of at least one of the major manifestations with two of the minor manifestations (fever, abdominal or præcordial pain, erythema marginatum, epistaxis, pulmonary changes and laboratory abnormalities).

- 3 The presence of rheumatic heart disease increases the diagnostic significance of the minor manifestations, when no other cause for these manifestations exist.

“Small, though probably insignificant, errors may be found with these criteria. Numerous clinical entities as enumerated may be confused with rheumatic fever. Clinical observations and, wherever possible specific diagnostic tests should be applied in any diagnostic problem.”

There are three other experiences in connection with rheumatic heart disease that are worthy of special mention in this lecture. The first concerns the treatment of the active process by the salicylates and the hormones, the second that of a change in severity of the arthritis since the 1920's, third, the treatment of chronic rheumatic valvular disease by surgery.

In the spring of 1918, when I served as internist and cardiologist at U.S. Base Hospital No. 6 (the Massachusetts General Base Hospital Unit) of the A.E.F. at Talence, near Bordeaux, there was an epidemic of streptococcus sore throat among the American troops stationed in south-western France. Shortly afterwards a convoy of about six dozen soldiers, acutely ill with rheumatic fever, came to the hospital. Dr. Richard Cabot, Chief of the Medical Service, then suggested that we try an interesting experiment, which we did. Half of these soldiers were put in one ward and treated with massive doses of the salicylates, chiefly in the form of aspirin. The other half of the cases were put into another ward and treated with analgesics and narcotics. Those who received aspirin were, within 24 to 36 hours, made completely comfortable with reduction of fever, while those treated with pain relieving drugs of other sort, continued to be miserable although sedated. Their pains were not adequately relieved and they continued to be febrile. It was impossible to maintain this experiment for more than a few days, because it was so evident that the salicylates were at least semi-specific in their effect. We almost thought that they were curative, but, of course, the active process itself and the heart disease were not completely relieved, even though the symptoms were.

A few decades later when the hormones, ACTH and cortisone, were introduced and were hailed with enthusiasm as curative for many conditions including rheumatic fever, these experiments were reinstituted. All of you know of the co-operative study carried out a few years ago in Britain and the U.S.A., which demonstrated quite clearly that both the salicylates and the hormones have a favourable effect on the rheumatic activity, although not specific enough to be considered as cures. There has been some dispute since as to greater specificity of the hormones. From observation of cases at the House of the Good Samaritan I have the belief that the hormones are more specific than salicylates, but they can sometimes have unfortunate secondary effects. Dr. Massell, of the House of the Good Samaritan, believes, from his experience, that there has been some definite saving of lives of youngsters with very fulminating rheumatic fever, through the use of the hormones. We still need something better than either drug in the treatment of the acute process and, more important still, we need some

specific therapy that will interrupt the chain of events from the time of the onset of a streptococcus sore throat up to the onset of rheumatic fever.

The second subject mentioned above is that of the change that seems to have come in the last generation in the severity of the active rheumatic process itself. Perhaps there is some tendency to exaggerate the findings in the "good old days" when we used to have more snow, bigger blizzards, and larger hail stones. Many of us who are older can look back and remember youngsters with fulminating polyarticular rheumatism, so sick and uncomfortable that even moving the bedclothes produced acute pain. Just why there has been an amelioration of the active process, so far as the joints themselves are concerned, I do not know. It is true that aspirin is so universally used for any ache or pain that that may be the answer, but on the other hand the process itself may have become less active. Incidentally, it used to be thought that rheumatic activity and rheumatic fever were rare in the tropics and subtropics and this may well have been true so far as the fulminating process was concerned, but on careful study of many individuals who live in tropical or subtropical areas, for example, in the southern part of the U.S.A., or in Mexico, or in India, or in the Philippines, a lot of rheumatic heart disease is found. Very recently, when I was in the Near East, I found there too, pure mitral stenosis suitable for surgical relief in patients who lived in the oasis at Damascus or in other similar places in those semi-tropical countries.

In closing this discussion of rheumatic heart disease in childhood, I want to bear witness and pay tribute to the magnificent pioneering of the cardiovascular surgeons of our day. I had the privilege, even before 1920, to be a fellow resident of that remarkably able young surgeon, Elliott Cutler, at the Massachusetts General Hospital in Boston. Later he became assistant to Harvey Cushing at the Peter Bent Brigham Hospital, and in the 1920's pioneered in efforts to relieve mitral stenosis surgically. This effort failed due to lack of adequate technique and anæsthesia of the day, but happily attempts were revived, and this time successfully, by thoracic surgeons who, during the Second World War, had rich experience in handling hearts and lungs of soldiers and officers who were wounded. This allowed a certain group of vigorous and able pioneers to attempt again relief of mitral stenosis surgically. As a result of this new attack during the last six to eight years, many thousands of cardiac patients crippled by pulmonary congestion from mitral stenosis have been so wonderfully benefitted, that the operation for "pure" mitral stenosis has become almost routine in many cities throughout the world.

On the other hand, the other valve commonly affected in rheumatic heart disease, namely the aortic, has presented a different story. Neither aortic regurgitation nor aortic stenosis has yet become routinely amenable to surgery, but forward steps have been made to change this dark picture of only a few years ago. One of the first of these was through the introduction by Hufnagel of his famous ball valve. This has helped a good many individuals, but it is only, of course, part of the answer. The valve, like other devices introduced as a foreign body, is not always safe, and it also only partially corrects the difficulty, namely that of the regurgitation in the lower half of the body; this does, however, remove about half of the extra work of the heart, and I have seen considerable reduction of

heart size and complete clearing of symptoms in some patients so treated. What we really need, of course, is the introduction of a proper valve or the repair of a damaged valve in its proper location. Thus, so far as aortic regurgitation is concerned, we are still groping, but we have much promise through the research work of many individuals studying to correct this difficulty.

Aortic stenosis has been attacked surgically now for quite a few years, but until the last year or so I have felt that the risk of the surgery was greater than the risk of not doing it, except in the case of a few young people with congenital aortic stenosis. During the last year, however, I have come to realize, as a result of the decrease in the risk of the operation and improvement of the results, that the time is coming, in fact is here now, when the risk of not doing the operation may be greater than the risk of the surgery itself. Just recently, that is, within a few weeks, a young man from Canada, aged 29, with calcareous congenital aortic stenosis, has been wonderfully helped by Dr. Harken in Boston. However, we have not really reached a satisfactory stage in the treatment of this condition.

Finally, we may hope that preventive measures applied to the candidates for the disease through their collagen tissue reaction to the hemolytic streptococcus, may radically reduce the need of cardiovascular surgery within the next generation. This is, of course, our ultimate aim.

CONGENITAL HEART DISEASE.

And now let us turn to the other important type of heart disease in childhood, namely congenital cardiovascular defects. In my medical student and hospital internship days, forty or more years ago, we did know of a few of the congenital deformities of the heart and great vessels. The best known was patency of the ductus arteriosus, but we were not aware of some of its complications. We also knew about the simple and actually less common type of the ventricular septal defect, which we called Roger's disease, the small calibre of which was usually well supported for many years, despite its intense murmur with thrill found characteristically at the left sternal border. The large defects which we now encounter so commonly must have been called something else, perhaps triloculate hearts. And while speaking of two and three chambered hearts we did know of their existence and found one now and again, but we could not diagnose them antemortem. We had, I think, heard of atrial septal defects discovered postmortem, but we had not yet reached the stage of their recognition as a clinical entity, although, during the First World War, Maude Abbott, and later Lutembacher, recognized the combination of mitral stenosis, and an atrial septal defect called after the latter observer.

Cyanotic congenital heart disease, the *morbus cæruleus* or *maladie bleue*, was, of course, in those earlier years of the present century recognized and called as such. It came to be known generally as due in the great majority of cases as the tetralogy of Fallot—the combination of pulmonary stenosis, ventricular septal defect, dextroposition of the aorta, and the right ventricular hypertrophy. Fallot described the condition in Marseilles in 1888, but it had been well delineated already 111 years earlier in 1777 by Sandifort. Such is a common story of so-

called priority and the attachment of names to clinical syndromes and pathological entities. In time other congenital cardiovascular causes of cyanosis were named, for example, the combination of a high ventricular septal defect with overriding of the aorta but no pulmonary stenosis. This was called Eisenmenger's syndrome, but it is, of course, only one variety of a ventricular septal defect. Then there was the so-called triology of Fallot with pulmonary stenosis, a large right ventricle, and a large atrial septal defect—a patient of mine with this condition, proved postmortem, lived to be 74 years old. A rare case of a tri- or biloculate heart with cyanosis would be encountered, but we did not recognize, at least at all clearly, in those early days, cases with cyanosis due to a reversal of shunt which commonly developed months or years after, and not at, birth. Examples of this now well known, of course, are instances of large patent ducti of Botalli and atrial septal defects with pulmonary hypertension.

Finally, uncomplicated pulmonary stenosis, though sometimes diagnosed, was considered rare in contrast to our current experience, while congenital aortic stenosis was practically always called rheumatic or calcareous. We did not know anything clinically about coarctation of the aorta, tricuspid atresia, Ebstein's anomaly of the tricuspid valve, pulmonary veins draining into the right atrium, a left sided or double superior vena cava, a congenital aneurysm of a sinus of Valsalva, anomalies of the coronary arteries, or a common arterial trunk.

Before leaving a general discussion of congenital heart disease, I would like to present one more quotation from my Discourse presented on May 21st, a month ago, at the annual meeting of the Massachusetts Medical Society in Boston. It was entitled *Genes, The Heart, and Destiny* (White 1957).

“By so-called congenital heart disease we really mean both truly inherited defects and those acquired in utero. There are no satisfactory terms to indicate this in common use today, but they should be introduced as soon as we are able to distinguish between the two. Both groups together might better be called ante-natal rather than congenital; those really inherited in the genes of the germ plasm might be eventually labelled ‘hereditary,’ or ‘inherited,’ or ‘intrinsic congenital.’ By strict definition ‘congenital’ should be the term to apply. The defects acquired during foetal life might be called ‘acquired in utero, or foetal,’ or ‘extrinsic congenital’ or even ‘connate’ as suggested by the dictionary. At present, however, we know next to nothing about the ætiologic factors actually behind either group or how to distinguish between them, and, indeed, there may be a mixture of the two, even when German measles in the first three months of pregnancy is responsible. In such a case it is conceivable that an inherited resistance to German measles or its lack, may be just as responsible for its occurrence, as exposure to the infection itself. In laboratory animals various other causative factors, such as vitamin deficiencies, anoxia and exposure to excessive radioactivity, are being tested and studied, but there is a great deal still to be learned.”

Now, in the last part of my lecture, let me present some of the current thoughts and experiences of myself and of my colleagues about a few of the more

important congenital defects of the heart and aorta; beginning with one of the oldest and best known, namely *patency of the ductus arteriosus*.

In the old days, patency of the ductus arteriosus was, for most of us, a simple condition for which we could do nothing except give common sense advice. We rarely saw long survival, that is, into old age, although there have been exceptions. In a paper which I presented at the Pædiatric Research Conference on Congenital Heart at the University of California Medical Centre in Los Angeles, in the fall of 1954 (White 1954), I referred to my own series of nearly one hundred private cases as follows :—

“My first private case, on December 19, 1920, was a woman of 64 who was ill with pneumonia; she lived until April, 1922, when she died of heart failure; autopsy showed a rather narrow lumen in the ductus and a slight to moderate degree of calcareous aortic stenosis. Several other of my patients with this condition have died of left ventricular failure—a woman died in 1925 after childbirth, at age 31; another succumbed on the operating table during the early days of surgery for this condition, at age 26. Several patients have had subacute bacterial endocarditis, generally fatal before the advent of antibiotics. One young man of 25, however, was cured in 1941 by ligation of the ductus. Confirmation of the cure in this case was obtained by autopsy five months later, following death in an automobile accident. Twenty-eight cases have been operated on successfully, although one ductus recanalized and required re-operation.

“At least five of my patients, first seen over 25 years ago, are in good health today; they are four men, now aged 58, 52, 65, and 78 years, and one woman aged 64. One boy, whom I first saw when he was 5 years old in 1928, had a murmur characteristic of ductus patency, but was perfectly well with no evidence of any cardiac abnormality at the age of 28.”

Two other points of importance about this deformity in our experience are: (1) that although we do see some long survivors, and although there may be no symptoms of trouble whatsoever right into middle age, nevertheless I always advise surgical correction now whenever I see such a patient, unless the patient is very old or obviously too sick to operate upon. (2) The next consideration is that of the reversal of shunt. I used to wonder whether or not a thrombosis in the ductus arteriosus might explain any clearing up or change of murmur. This is still a possibility, but the more common cause, which is itself nevertheless rare, is the neutralization of pressures in the pulmonary artery and aorta due to pulmonary hypertension secondary to pulmonary artery sclerosis. Usually there is not a fast flow of blood from the pulmonary artery into the aorta, but there is enough to give rise to cyanosis in the lower part of the body. This we did not know anything about or pay any attention to a generation ago. In such cases, of course, obliteration of the patent ductus is contraindicated.

Ventricular Septal Defect. An even more intriguing congenital deformity which we used to regard as simple, is the ventricular septal defect. This for us, years ago, was synonymous with the *maladie de Roger*, but with experience in the last

two or three decades from study of ventricular septal defects at autopsy, at operation, and by cardiac catheterization, makes it evident that actually Roger's disease includes the minority of cases with a ventricular septal defect. Since this is so and the ventricular septal defects that are more important clinically are larger and higher and may even involve the mitral valve, we have now come to regard the condition as much more serious and demanding surgical correction, which, happily, is beginning to be applied. Within the past two weeks I have seen a patient, in the Cardiac Clinic at the University of Minnesota, much limited by such a septal defect, and plans were being made to operate using the pump oxygenator. It has become a procedure almost routine there, although not many cases have as yet been done. While in Minneapolis I was told that Dr. Lillehei of Minneapolis was about to tour Europe for the purpose of lecturing on and actually demonstrating his technique for such operations.

These cases, as I have already noted, may or may not have cyanosis. Most of them do not, but if the aorta is somewhat dextroposed over the septal defect, then cyanosis can occur without pulmonary stenosis. At any rate, there is a strain on both ventricles, usually more on the right side, and the heart can be quite enlarged with failure or bacterial endocarditis threatening.

Atrial Septal Defects. An atrial septal defect I did not diagnose at all until 1933 in my second five thousand private cardiac cases. Not once in the first five thousand patients did I make such a diagnosis, so that this is a new clinical entity to most of us. Generally it is quite easy to diagnose with a much enlarged right ventricle and the characteristic great fullness of the pulmonary arc and lung hilus shadows. However, many patients continue surprisingly well and active, right into middle age, but very few survive into old age. Therefore, again surgical correction with the new techniques is in order and is being successfully carried out either by refrigeration, now improved by Brock's technique of cooling the blood rather than the patient, or by the use of the pump oxygenator. Until recently, however, surgical operations were done rather blindly. Now they can be carried out under direct vision with these new techniques of preparation. Excellent results are already accumulating.

Tetralogy of Fallot. The tetralogy of Fallot, which we knew about a good many years ago but which we sometimes called simply pulmonary stenosis in the early days, has been treated surgically and successfully by several techniques. The first was that of Blalock and Taussig by vascular anastomosis shunting blood back into the lungs from the systemic circulation, the second accomplishing the same result by Potts through direct connection of the aorta with a pulmonary artery, and the third, which is probably the simplest and now most popular, is that of Brock, by correcting the pulmonary valve or infundibular stenosis itself. One of the difficulties of the first two techniques is that one is introducing another factor of strain, that is, an additional shunt. Many of the patients who have been operated upon during the last ten years have done well, but it is still too early to prophesy finally about the ultimate success of any of the procedures. Eventually more complete correction of the difficulties with the new techniques may be envisioned.

Pulmonary Stenosis. Little need be said about uncomplicated congenital pulmonary stenosis, except that it is much more common than we used to think. Often, however, it is of such slight degree that there is no great burden for the heart, and life can continue into old age. Sometimes this seems to be more a physiological diagnosis than an anatomical one, in other words, there are slight degrees of pulmonary stenosis which are hardly evident, even at autopsy. This has been found by cardiac catheter studies. Severe grades, however, involving either the pulmonary valve itself or the infundibulum do need correction by Brock's technique.

Aortic Stenosis. Congenital aortic stenosis did not enter our experience until the 1930's. My first diagnosis of congenital aortic stenosis was in November, 1934. In the past, undoubtedly, as stated by Campbell, congenital aortic stenosis has been confused with rheumatic and calcareous aortic stenosis. It is probably not actually a rare condition. It too, may be well supported for a good many years, but eventually, if of high degree, the strain on the left ventricle becomes too great and left ventricular failure and death can result in middle age or indeed even in youth. Happily, as I have already mentioned under rheumatic heart disease, surgery is becoming more practical for aortic stenosis, especially of the congenital type, even though the valve is calcified. Of course, the ideal procedure would be to replace the damaged valve by a new one. This remains for the future.

Coarctation of the Aorta. Only once in my first five thousand cases, and that was toward the end of the series, did I diagnose coarctation of the aorta. It was a new clinical entity in the middle of my career. I diagnosed my first case in February, 1933, in a law student 23 years old. As in his case, hypertension in youth has been, in the majority of cases, due to this congenital defect, and this became evident, especially during the Second World War. However, until Crafoord and Gross independently introduced surgical correction of the condition in 1945, we had no treatment for it except limitation of activity and common sense advice otherwise. One of the early difficulties with the surgical approach was the finding of occasional cases with too long an area of coarctation or an additional defect, such as an aortic aneurysm below the coarctation that required replacement by a long blood vessel graft. Therefore, it was only when blood vessel banks came into use that we could feel confident about the correction of coarctation of the aorta in every case. Incidentally, we have found that it can be the site of bacterial endarteritis. As in the case of mitral stenosis and patency of the ductus arteriosus, coarctation of aorta has now become a condition to be routinely cured by surgery unless the patient is very old, or for other equally important reasons.

In closing this second part of my address, I would like to repeat the brief conclusions of my paper presented at the University of California Medical Centre at Los Angeles. They are as follows :—

“The clinician, pathologist, experimental embryologist, and surgeon, all encounter a confusing variety of congenital cardiovascular anomalies. Out of the chaos of a generation ago, however, has come some order through

the efforts of many workers. Marvellous advances have been effected in diagnosis, cure and palliation, but the solution of the main problem still eludes us.

"I would suggest three thoughts. First, there is every degree of almost every defect, and the prognosis is often largely related to this difference of the degree of trouble. For example, if a ductus arteriosus is very large, the future of the patient is bleak without speedy correction, while if of small calibre, it may allow a long, active life. One of my patients in this latter group still runs a large business and plays golf without symptoms, at the age of 78 years.

"Second, there are combinations of defects, or indeed degrees of a single defect, that are incompatible with life. If we include those cases for whom we can do nothing in our clinical statistics, we should be inclined to be more humble.

"And third, we should spend more time, clinically and experimentally, in trying to understand the pathogenesis of these defects. An ounce of prevention will be here, as elsewhere, worth a pound of cure."

This brief survey of my own experience with heart disease in childhood, has, I hope, been of interest to you, not only because of its historical aspect, but because here and there I have presented some current viewpoints about diagnosis, treatment, and prevention, which may be of practical value to you. Thanks very much for listening to me.

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